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Budd-Chiari Syndrome Due To Echinococcus Multilocularis Infection: A Case Report

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Abstract:
Hepatic vein obstruction is associated to a clinical and pathological presentation named the Budd-Chiari syndrome, including hepatic sinusoidal dilatation and congestion, ascites and abdominal pain, as a result of hepatic outflow block.
Echinococcal disease is caused by infection with the metacestode stage of the tapeworm Echinococcus. The most common presenting symptoms include malaise, weight loss, and right upper quadrant discomfort due to hepatomegaly.
Hepatic vein invasion, typically associated to Echinococcus multilocularis has rarely been described .We provide here a detailed observation of a acute Budd-Chiari syndrome due to Echinococcus multilocularis infection. Detection of Budd-Chiari syndrome in case of Echinococcus multilocularis infection is important for the appropriate treatment.

Key words: Echinococcus multilocularis, Budd-Chiari Syndrome.

The radiological evidence of hepatic vein thrombosis, enlarged liver, increased abdominal pain, congestion around the central vein of the liver lobule at histology, elevated portal pressure, raised serum aminotransferases and presence of ascites rendered the diagnosis of the Budd-Chiari syndrome (BCS) most likely.[1]

Adult form infections of Echinococcus multilocularis (EM) are perpetuated in a sylvatic cycle with wild carnivores—mainly red (Vulpes vulpes) and arctic (Alopex lagopis) foxes—regarded as the most important definitive hosts. Domestic dogs and cats can also harbour the tapeworm and may be involved in a synanthropic cycle. Small mammals (usually microtine and arvicolid rodents) act as intermediate hosts. The metacestode of EM is a tumour-like, infiltrating structure consisting of many small vesicles embedded in stroma of connective tissue.[2]

Although many disorders including parasitic diseases can cause BCS. EM compressing hepatic veins leading to acute BCS has been rarely reported in English medical literature. This case report outlines the approach to clinical diagnosis in patients who have BCS. So, we report a case of BCS due to EM.

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Case Presentation

15-year-old girl was admitted to our hospital because of malaise, weight loss, and right upper quadrant discomfort. She had been well until five weeks earlier. The patient was not sexually active and she took no medications. She resided with her parents and five siblings in a village.

Examinations of the head, neck, and chest and cardiovascular systems showed no abnormalities. On abdominal examination there was moderate tenderness in the right upper quadrant without rebound or guarding and there was mild ascites. A palpable liver edge 3 cm below the right costal margin was detected ;and the splenic tip was palpable. Abdominal ultrasonography showed huge cystic, calcified and heterogenous lesions in the right lobe of the liver and splenomegaly and ascites. Abdominal paracentesis was performed and the serum– ascites albumin gradient was reported to be 1.3 g/dl. The total protein level was 10.3 g/dl, the globulin level was 6.9 g/dl, the alkaline phosphatase level was 554 U/l, the gamma glutamyl transferase level was 110 U/l. The white-cell count was 18,800 X 10^9/l, with 65 % neutrophils and 20 % eosinophils. Total Ig E level was 22.400 U/l. An ELISA test for EM was positive.

Abdominal CT scan demonstrated nonenhanced, calcified,centrally necrotic, irregular areas in the right lobe of the liver . The right lobe was measured 11x 10 cm. and there were hepatic and right portal vein invasion. The caudate lobe was hypertrophic and the

paranchimal enhancement was seen in massive pattern (Figure1).

Reported CT findings were compatible with BCS due to EM. Albendazole was given at a dose of 10 mg/kg/day and surgical resection was planned.

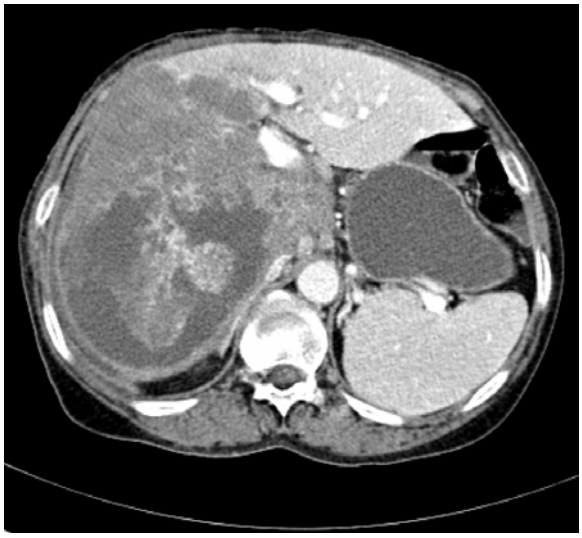

Figure I. CT scan of abdomen; heterogeneous hypodense and pseudocystic necrotic areas, calcifications, and enlarged caudate lobe and invased hepatic and right portal vein.

Discussion

This 15-year-old girl was in good health until five weeks before admission, when abdominal distention and discomfort due to ascites and hepatosplenomegaly developed. A typical feature of this case is the high gradient ascitic fluid so the differential diagnosis is narrowed to causes associated with high serum–ascites albumin gradient (≥1. 1 g/dl) ascites. These are hepatic cirrhosis, alcoholic hepatitis, hepatic outflow obstruction (BCS and veno-occlusive disease) and cardiac ascites.[3] The absence of a history of stem-cell transplantation effectively rules out veno-occlusive disease, and the absence of cardiac findings such as regurgitant murmur, jugular venous distention, pericardial knock, or pulsatile liver on physical examination rules out cardiac causes. Furthermore; there were no alcohol and herbal medicine consumptions and no cirrhosis releated findings.The clinical course of this girl's illness, with the rapid development of liver disease over a period of a few weeks, is typical of the BCS. Abdominal pain, hepatomegaly, and ascites are present in almost all patients with the Budd–Chiari syndrome. Furthermore; a parasitary disease kept in mind because of elevated levels of globulin, eosinophils and total IgE. All these findings were presented in our case.[4] In addition to above findings; an enlarged caudate lobe, as was seen in this girl, was supported the diagnosis of the BCS.

The BCS is a heterogeneous group of disorders characterized by hepatic venous outflow obstruction at the level of the hepatic venules, the large hepatic veins, the inferior vena cava, or the right atrium.[5] (Table-1).

Table I. Causes of the Budd–Chiari Syndrome.

1-Myeloproliferative disorders (50% of cases) (Polycythemia vera, essential thrombocythemia)
2-Cancer (Hepatocellular, adrenal, renal carcinomas: direct spread or production of erythropoietin)
3-Hypercoagulable states (Factor V Leiden, prothrombin gene mutation G20210A, antiphospholipid antibody, antithrombin III deficiency, protein C deficiency, protein S deficiency, paroxysmal nocturnal hemoglobinuria)
4-Oral contraceptive pills or pregnancy
5-Behçet's disease
6-Hepatic infections (abscess, echinococcal cyst)
7-Membranous webs of inferior vena cava (congenital or acquired)
8-Idiopathic (up to 20% of cases) (Occult myeloproliferative disorder;*JAK2* mutations)

Factors that confer a predisposition to the development of the BCS, including hypercoagulable states, both hereditary and acquired, and a variety of other causes, can be identified in about 75 percent of patients [6]

Hematologic abnormalities,particularly myeloproliferative disorders, are the most common causes of the Budd–Chiari syndrome. Polycythemia vera accounts for between 10 and 40 percent of cases of the syndrome,[7] whereas essential thrombocythemia and myelofibrosis are less prevalent causes. Endogenous erythroid-colony formation may be seen in up to 87 percent of patients thought to have idiopathic BCS, suggesting that the majority of patients in whom the cause of the BCS is not apparent have a myeloproliferative disorder.[8] Neither of these haemalotogic disorders is overtly evident in this patient.

Serum aspartate and alanine aminotransferase levels may be more than five times the upper limit of the normal range in the fulminant and acute forms of

the BCS, whereas increases are smaller in the subacute form. Serum alkaline phosphatase and bilirubin levels also increase to a varying extent, along with a decrease in serum albumin as was seen in this case. [9] The BCS due to hepatic infections (abscess, echinococcal cyst) is very rare (Table 1) Reported CT findings of liver infestation by EM include heterogeneous hypodense areas with lack of contrast enhancement, isodense areas with or without contrast enhancement, calcifications, pseudocystic necrotic areas and dilatation of intrahepatic bile ducts.[10] CT patterns of EA lesions can be quite similar to those seen in primary hepatic neoplasms and metastases.[11] On CT, the lack of contrast enhancement in a hypodense lesion with calcifications was helpful in differentiating EM from liver neoplasms.All the radiologic findings was compatible with EM. Thus, the BCS in this case was explained by EM.

Cases of EM are characterised by an initial asymptomatic incubation period of 5–15 years, and a subsequent chronic course. The right lobe is involved most frequently, with involvement of the porta hepatis or multiple lobes being less frequent. Parasitic lesions in the liver can vary from small foci a few millimetres in size to large (15–20 cm in diameter) areas of infiltration. Extrahepatic primary disease is very rare.(1% of cases).[12,13] One-third of cases present with cholestatic jaundice, one-third present with epigastric pain, and the remainder present with vague symptoms like weight loss or fatigue, or are noted to have incidental hepatomegaly.[12] Our patient was presented with most of these symptoms.

The definitive diagnosis for most cases of EM is established by physical imaging methods, such as radiology, ultrasonography, computed axial tomography (CT scanning), and magnetic resonance imaging.[14]

Immunodiagnosis is useful not only in primary diagnosis but also for follow-up of patients after surgical or pharmacological treatment.[15] ELISA, indirect haemagglutination antibody assay, latexagglutination test, and immunoblot test are the most commonly used immunological methods..[16] ELISA results of our case revealed current infection with EM.

The diagnosis of EM is based on similar findings and criteria including case history, clinical findings, morphological lesions identified by imaging techniques and immunodiagnosis as in our case.

Radical surgery—as for hepatic malignancy—has been the historical cornerstone of treatment for alveolar echinococcosis.[17]

Albendazole is only parasitostatic against the EM metacestode. Chemotherapy should be continued for 2 years after surgery. The role of life-long chemosuppression is being explored. Liver transplantation has been undertaken in some patients with alveolar echinococcosis.[18]

Therapy for patients with the BCS includes medical management and the relief of hepatic venous outflow tract obstruction in order to prevent necrosis, with liver transplantation in selected patients, especially those with fulminant hepatic failure.[9]

In this case, surgical resection of the involved hepatic segment may be require because of the complicating BCS and it may therefore be reasonable to recommend long-term chemotherapy such as albendazole after hepatic resection. Human EM is a potentially fatal, chronically progressive hepatic infestation. There is a risk of the BCS. Therefore, we suggest that EM should be considered in the differential diagnosis of the BCS especially in the rural areas.

We reports a rare case of BCS in a patient with EM infection and we would like to emphasize the role of rare parasitary diseases such as EM in the BCS.

Echinococcus Multilocularis İnfeksiyonuna Bağlı Budd-Chiari Sendromu: Bir Olgu Sunumu

Özet:

Hepatik ven obstruksiyonu; Budd- Chiari Sendromu olarak adlandırılan ve hepatik sinuzoidal dilatasyon, konjesyon, asit ve karın ağrısına neden olan klinik ve patolojik bir durumdur.

Ekinokokkal hastalık ekinokokun metasestod evresiyle infeksiyon sonucu gelişir. En sık başvuru semptomları; halsizlik, kilo kaybı ve hepatomegaliye bağlı sağ üst kadran ağrısıdır. Echinococcus multilocularis infeksiyonuna bağlı Hepatik ven invazyonu nadiren tanımlanmıştır. Bizler burada E. Multilocularis infeksiyonuna bağlı Budd – Chiari sendromunu tanımlıyoruz. Bu infeksiyonun seyrinde Budd Chiari sendromunun tesbit edilmesi uygun tedavi için önemlidir.

Anahtar kelimeler: Echinococcus multilocularis infeksiyonu, Budd-Chiari Sendromu.

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